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LETTER TO THE EDITOR

Value of CMR to Differentiate Cardiac Angiosarcoma From Cardiac Lymphoma



Primary cardiac malignancies are extremely rare (1). The 2 most common tumors (i.e., angiosarcoma and lymphoma) are usually right sided and often located near the right atrial wall and right atrioventricular groove, hampering differentiation on cardiac imaging. We describe the value of cardiac magnetic resonance (CMR) in 12 patients with these tumors (7 angiosarcomas and 5 cardiac lymphomas [all B cell non-Hodgkin, HIV negative], all confirmed by biopsy [n = 11] or autopsy [n = 1]). Patients with angiosarcoma were younger than patients with lymphoma,

and dyspnea was the most common complaint at initial presentation for both tumors. Maximal tumor diameter was similar between the tumors (84 ± 22 mm for angiosarcoma vs. 88 ± 19 mm for lymphoma). The right atrial wall was partially to completely invaded by tumor in all patients (Figure 1A to 1F, Online Video 1). The right atrial appendage was involved in all patients with an angiosarcoma but in only 1 patient with lymphoma ($p = 0.01$) (Online Video 2). In contrast, all lymphomas diffusely involved the right atrioventricular groove and completely surrounded the right coronary artery (defined as >10 mm tumor tissue around the entire circumference) (Figures 1D and 1E). The angiosarcomas showed partial involvement of the right atrioventricular groove in 4 patients and only incompletely ($<180^\circ$) encased the right coronary artery (RCA) ($p < 0.001$) (Figure 1A). Extension toward the right ventricle was present in all lymphomas but

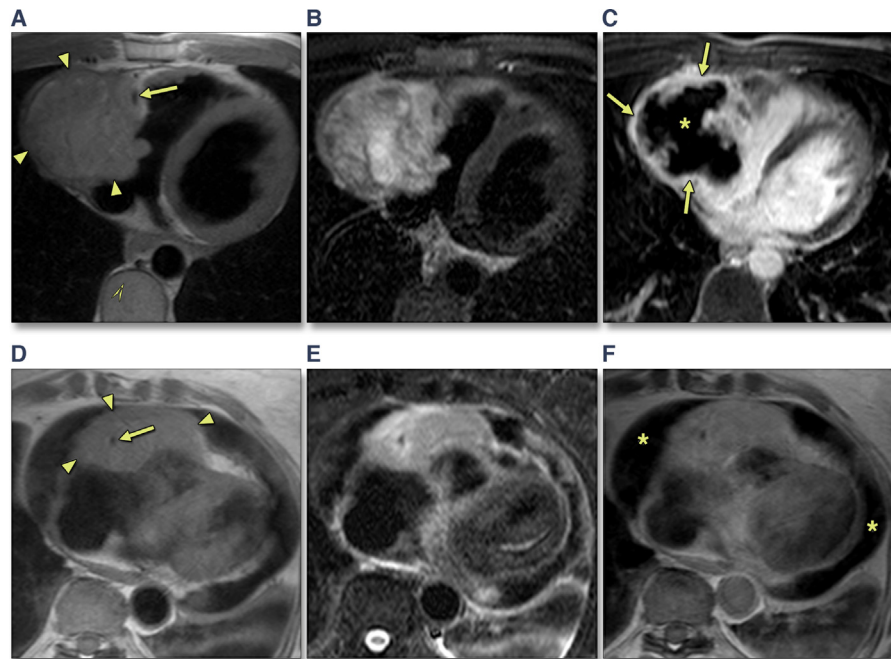


FIGURE 1 Representative Examples of Cardiac Angiosarcoma and Cardiac Lymphoma

(A to C) Cardiac angiosarcoma. (D to F) Cardiac lymphoma. Axial T1-weighted fast-spin-echo cardiac magnetic resonance (CMR) (A and D). Axial T2-weighted short-tau inversion-recovery fast-spin-echo CMR (B and E). Axial late gadolinium enhancement (LGE) CMR (C) and T1-weighted fast-spin-echo CMR post-contrast (F). The angiosarcoma presents as a voluminous mass (A, arrowheads) arising from the right atrial wall extending to the pericardium, right atrial cavity, and right coronary artery (A, arrow) with strong rim enhancement (C, arrows) and central liquefaction (C, asterisk) on LGE CMR (Online Video 1). The cardiac lymphoma presents as a mass mainly centered in the right atrioventricular groove (D, arrowheads), diffusely surrounding the right coronary artery (D, arrow), extending to the right atrial and right ventricular wall (Online Video 2). Homogeneous enhancement post-contrast administration (F). Also note the presence of a circumferential pericardial effusion (F, asterisk).

only in 3 of 7 angiosarcomas (Figures 1D and 1E). The tricuspid valve was involved occasionally and was no different between groups. The CMR images showed a central liquefaction necrosis (or necrotic areas) in 6 of 7 angiosarcomas (i.e., rim enhancement with lack of central enhancement) (Figure 1C). This pattern was absent in all lymphomas that showed homogeneous tumor enhancement ($p = 0.015$) (Figure 1F). Pericardial effusion was common in both conditions (5 of 7 angiosarcomas [range 10 to 30 mm] and 4 of 5 lymphomas [range 27 to 45 mm]) (Figures 1D and 1F). Except in 1 patient with angiosarcoma, enhancement of the pericardial layers was shown on post-contrast CMR images in all patients. Pericardial “nodular” metastasis (including the transverse/oblique sinus) was present about half of the time in both conditions, whereas pulmonary metastases were found only in angiosarcomas (3 of 7 patients).

The current results obtained in a small but representative group of patients with cardiac angiosarcoma and HIV-negative lymphoma showed that CMR can help differentiate between these malignant tumors. Tumor location in the right atrial wall and atrial appendage in a younger patient with central necrosis and possibly pulmonary metastasis favors angiosarcoma, whereas lymphomas appear in older patients with diffuse right atrioventricular involvement and RCA encasement and as solid tumors on CMR imaging. Although this is a small sample and

features may depend on when the diagnosis was made in the natural history of the disease, it represents a significant number of biopsy-proven tumors with CMR correlation in these rare conditions. CMR may help noninvasive differentiation between angiosarcoma and lymphoma.

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APPENDIX For accompanying videos and their legends, please see the online version of this article.